# Healthcare Burden Among Patients with Chronic Inflammatory Demyelinating Polyneuropathy (CIDP): A Targeted Literature Review (TLR)

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## INTRODUCTION

- Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic, progressive or relapsing-remitting autoimmune-mediated neurological disease that impacts the peripheral nervous system, manifesting as sensory loss and muscle weakness in the extremities, which may lead to long-term disability and significant limitations in activities of daily living.[1,2]
- Current guidelines recommend intravenous immunoglobulin (IVIg), plasma exchange, and corticosteroids as first-line (1L) treatment, subcutaneous immunoglobulin as maintenance treatment, and immunosuppressive agents as other treatment options.[1]
- Novel treatments that target specific immune cells or signaling mechanisms involved in the pathogenesis of CIDP are emerging as alternative treatment options, including the recent approval of a neonatal Fc receptor blocker.[2-5]

## OBJECTIVES

- **To evaluate the burden of CIDP, including clinical, humanistic, economic, and treatment-related aspects, through an assessment of the scientific literature.**

## METHODS

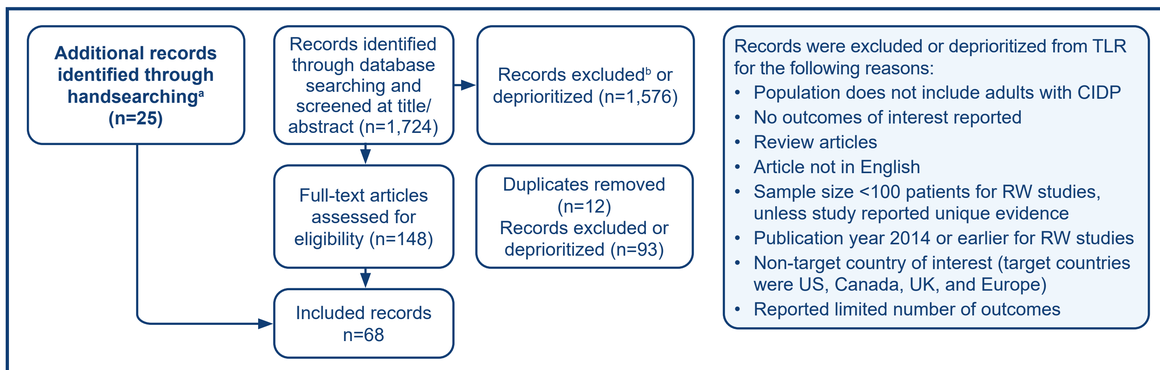
- Searches were conducted in March 2025 for articles from Embase and MEDLINE/MEDLINE In-Process using a pre-defined search strategy (no date restriction) and supplemental searches of health technology assessments, primary sources from review articles, and relevant conference abstracts (published January 1, 2021 onwards).
- Regions of interest were US, Canada, UK, and Europe, with other countries included if key gaps were addressed.
- One independent reviewer screened records with the assistance of an artificial intelligence screening tool (Nested Knowledge), first as titles/abstracts and then as full texts; a second independent reviewer performed a quality control check of 10% of records.

## RESULTS

### Study attrition

- From 1,749 unique records evaluated, 68 articles or conference abstracts were included. (**Figure 1**)

**Figure 1. Identification of Source Materials**

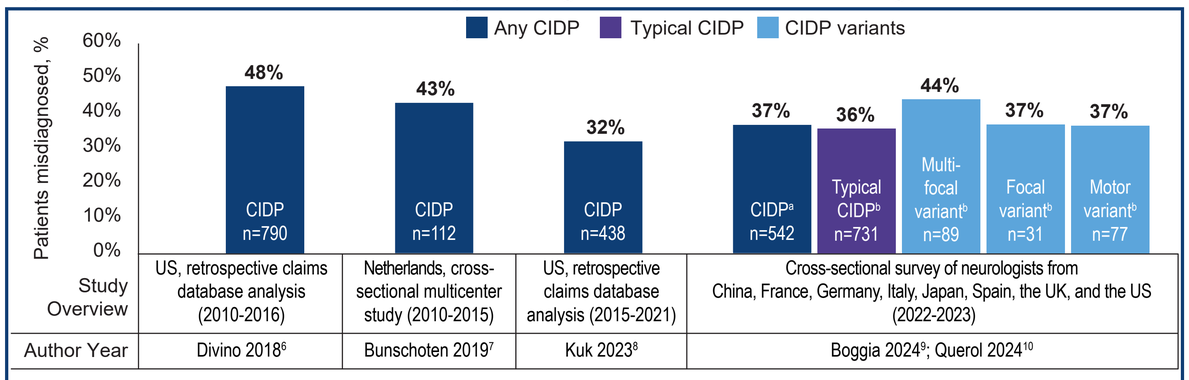



[a]Conference abstracts from select conferences of interest not indexed in Embase and primary sources identified from review articles were hand-searched per the protocol. [b]1,145 records were excluded using a trained AI screening model per the protocol.
Abbreviations: AI = artificial intelligence; CIDP = chronic inflammatory demyelinating polyneuropathy; RW = real-world; TLR = targeted literature review

### Diagnosis patterns

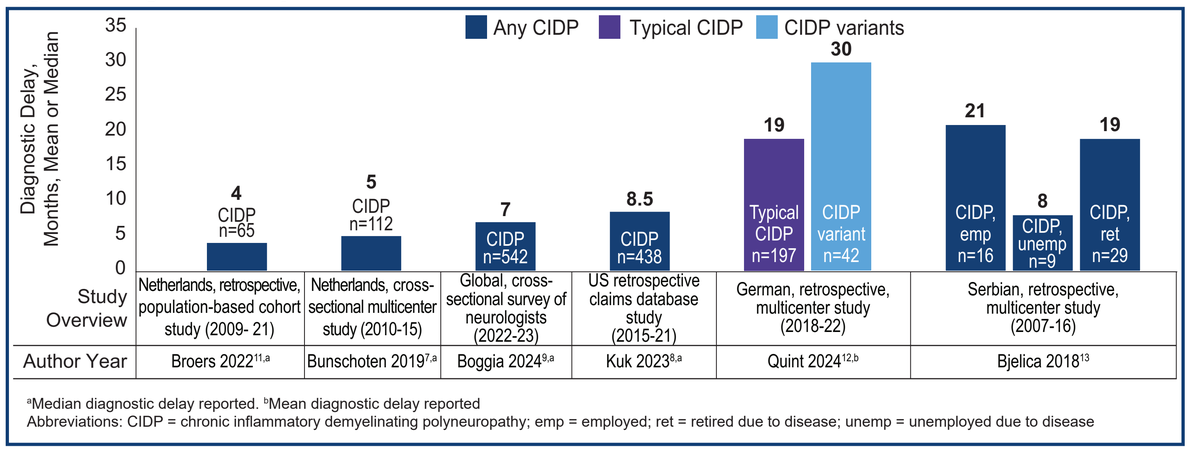
- Approximately 30% to 50% of patients with CIDP are initially misdiagnosed (**Figure 2**)[6-10] and can experience a 4- to 30-month diagnostic delay after symptom onset (**Figure 3**).[7-9,11-13]

**Figure 2. Proportion of Patients Receiving a Misdiagnosis Prior to the Correct Diagnosis of CIDP**




[a]Data provided by 83 physicians for 542 total patients. [b]Data provided by 164 physicians for 1,056 total patients
Abbreviations: CIDP = chronic inflammatory demyelinating polyneuropathy

**Figure 3. Diagnostic Delay in Patients With CIDP**




[a]Median diagnostic delay reported. [b]Mean diagnostic delay reported
Abbreviations: CIDP = chronic inflammatory demyelinating polyneuropathy; emp = employed; ret = retired due to disease; unemp = unemployed due to disease

- Patients with a longer time from symptom onset to diagnosis or treatment for CIDP had worse outcomes than those with earlier diagnoses or treatment initiation.
  - In a Dutch cross-sectional study (2010 to 2015; n=112), patients diagnosed >5 months vs. ≤5 months after symptom onset had significantly worse fatigue (Rasch-built Fatigue Severity Scale; *P*=0.036) and disability (Rasch-built Overall Disability Scale; *P*=0.008) at study entry (i.e., after a median CIDP disease duration of 9 years).[7]
  - In a German retrospective analysis (2018 to 2022; n=197), patients with a delay of >12 months vs. ≤12 months from symptom onset to treatment initiation experienced significant worsening in Inflammatory Neuropathy Cause and Treatment (INCAT) leg disability scores over a 36-month follow-up period after diagnosis (*P*<0.01).[12]
  - In a UK and Korean retrospective analysis (2014 to 2023; n=144), patients with a delay of >12 months vs. ≤12 months from symptom onset to treatment initiation had significantly worse disability at a median of 6 years of follow-up, as measured by INCAT (*P*=0.0026), Overall Neuropathy Limitation Scale (*P*=0.0018), and Inflammatory Rasch-built Overall Disability Scale (*P*=0.00064) scores.[14]

### Treatment patterns

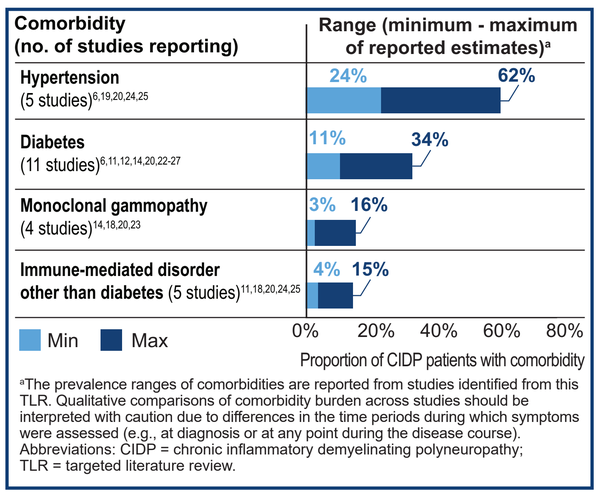
- The most common 1L treatments in the US and Europe were IVIg and corticosteroids.
  - In Europe, 1L IVIg use alone ranged from 53% to 80%, 1L corticosteroid use alone ranged from 12% to 39%, and combination therapy with IVIg and steroids ranged from 5% to 8%.[7,12,14,15]
  - In the US, 1L IVIg use alone ranged from 11% to 43%, 1L corticosteroid use alone ranged from 32% to 57%, and combination therapy with IVIg and steroids ranged from 8% to 12%.[6,16,17]
- Limited data on real-world treatment patterns for second-line (2L) or later therapy suggest that approximately one-quarter to two-thirds of patients who initiate 1L therapy for CIDP will switch to a different 2L monotherapy.[12,18]

## RESULTS (CONT.)

### Clinical burden

- Results from a German, prospective, noninterventional, multicenter study (2012 to 2016) demonstrated that patients with CIDP (N=148) treated with Gamunex 10% (IVIg) over a 96-week observational period had substantially lower physical and physical-role functioning Short Form 36 health survey scores compared with German general population norms (statistical comparison not provided).[19]
- Comorbidities (i.e., back pain, cerebrovascular disease, hypothyroidism, leukemia, neuropathic pain, osteoarthritis, peripheral vascular disease, and sleep apnea) were significantly more prevalent in patients with CIDP at time of diagnosis compared with matched controls in a US-based retrospective claims database analysis (2010 to 2016; N=790 patients with CIDP).[6]
- Common comorbidities reported in patients with CIDP are shown in **Figure 4**.
  - Long-term corticosteroid or immunoglobulin use can be associated with hypertension.[20,21]
  - Long-term corticosteroid use can be associated with diabetes.[20,21]

**Figure 4. Comorbidity Burden in Patients With CIDP[6,9,11,12,14,18,20,22-27]**




[a]The prevalence ranges of comorbidities are reported from studies identified from this TLR. Qualitative comparisons of comorbidity burden across studies should be interpreted with caution due to differences in the time periods during which symptoms were assessed (e.g., at diagnosis or at any point during the disease course).
Abbreviations: CIDP = chronic inflammatory demyelinating polyneuropathy; TLR = targeted literature review.

### Patient-reported Outcomes

- Patient dissatisfaction with treatments is associated with significantly worse disability and decreased quality of life (**Table 1**).[28,29]

**Table 1. Impact of Treatment Satisfaction on Disability and Quality of Life in Patients With CIDP**

| Study Overview | Summary of Results | | |
|---|---|---|---|
| US and Canadian online survey of patients with CIDP (N=318) from 2019 to 2020[28] | Patients who were dissatisfied with their treatment (n=83) vs. satisfied with their treatment (n=222) had significantly worse mean disability and quality of life scores (*P*<0.01): | | |
| | | **Dissatisfied** (n=83) | **Satisfied** (n=222) |
| | R-ODS[a] | **28** | **33.5** |
| | ONLS[b] | **4.2** | **3.5** |
| | INCAT[b] | **3.14** | **2.4** |
| | SF-6D utility[c] | **0.56** | **0.64** |
| | EQ-5D utility[c] | **0.33** | **0.54** |
| | EQ-5D VAS[c] | **52** | **61** |
| | CAPPRI[d] | **20.1** | **14.4** |
| Cross-sectional survey of patients with CIDP (N=318); study years not reported[29] | Dissatisfaction with treatment was found to be a main driver of worse health utility scores for the EQ-5D-5L and the SF-6D, based on a multivariable linear regression model (data not provided). | | |

[a]Lower scores indicate worse disability. [b]Higher scores indicate worse disability. [c]Lower scores indicate worse QoL/utility. [d]Higher scores indicate worse QoL.
Abbreviations: CAPPRI = Chronic Acquired Polyneuropathy Patient-Reported Index; CIDP = chronic inflammatory demyelinating polyneuropathy; EQ-5D = EuroQol 5 Dimension; INCAT = Inflammatory Neuropathy Cause and Treatment; ONLS = Overall Neuropathy Limitation Scale; R-ODS, Rasch Overall Disability Scale; SF-6D = Short-Form 6-Dimension; VAS = visual analog scale

- Reasons for treatment dissatisfaction and discontinuation:[30-32]

**Physicians**

**Dissatisfaction**:

- *"Declining efficacy over time"*

**Patients**

**Dissatisfaction**:

- *"Dosing frequency"*
- *"IV route of administration"*

**Patients**

**Discontinuation**:

- *IVIg was "not beneficial"*
- *Corticosteroids had "lack of benefit" and "poor tolerability"*

Abbreviations: IVIg = intravenous immunoglobulin

### Economic burden

- CIDP incurs substantial economic burden, including treatment-related costs, hospitalizations, and productivity loss (**Table 2**).[6,33-36]

**Table 2. Summary of Economic Burden of CIDP**

| Key Theme | Summary of Results |
|---|---|
| **Total Cost Burden** | **CIDP is associated with significant economic burden**<br>• Patients with CIDP incurred significantly greater total costs, inclusive of outpatient pharmacy and medical services costs, over a 2-year follow-up period compared with matched controls ($116,330 vs. $15,586 in 2016 USD; P<0.0001), based on a US retrospective claims database analysis.[6]<br>• Approximately two-thirds of patients with CIDP were hospitalized in the year following diagnosis, based on a Swedish population-based cohort study (2001 to 2017).[36] |
| **Treatment Costs** | **Treatment with Ig contributes substantially to total costs**<br>• Patients with CIDP treated with IVIg monotherapy had higher mean therapy costs from treatment initiation to end of a 2-year follow-up period compared with those treated with corticosteroid monotherapy ($165,000 vs. $7,900 in 2016 USD), based on a US retrospective claims database analysis.[6]<br>– Study authors noted that higher costs of IVIg may have been related to longer persistence on IVIg vs. corticosteroids; setting of care (e.g., inpatient, outpatient, or home-based) was not evaluated, and cost components were not reported.[7]<br>• Chronic Ig users had higher mean all-cause medical costs than intermittent Ig users ($206,418 vs. $64,722 PPPY), based on a US retrospective claims database analysis (2016 to 2020).[33]<br>– Components of all-cause medical costs, cost year, and type of Ig (SC or IV) were not reported. |
| **Disease Burden** | **Worsening impairment increases the economic burden associated with CIDP**<br>• Patients with severe impairment vs. mild impairment (per INCAT scores) experienced higher rates of hospitalization (4.5 vs. 1.5) and higher mean IVIg paid amount ($125,538 vs. $80,248) in the first year following diagnosis, based on a US retrospective claims database analysis.[34]<br>– Cost year and study period were not reported in the 2024 conference abstract.<br>• Depression and lower HRQoL were both significant predictors (P=0.033) of higher total costs, based on a German cross-sectional study (2013 to 2014).[35] |
| **Indirect Costs** | **Work interruption contributes substantially to the indirect costs associated with CIDP**<br>• Premature retirement was the second largest contributor to total costs among patients with CIDP after medication, based on a German cross-sectional study (2013 to 2014)[35]<br>• 60% of patients with "likely" CIDP either stopped work (44%) or changed the way they worked (16%), based on a global online survey (2017 to 2018).[32]<br>• The unemployment rate in CIDP patients was 18% (9% of patients attributing to CIDP), which was greater than the general Serbian population (13%) in 2017, based on a retrospective study.[13] |

Abbreviations: CIDP = chronic inflammatory demyelinating polyneuropathy; HRQoL = health-related quality of life; Ig = immunoglobulin; INCAT = Inflammatory Neuropathy Cause and Treatment; IVIg = intravenous immunoglobulin; PPPY = per patient per year; SC = subcutaneous; USD = US dollar

## CONCLUSIONS

- **Significant health-related quality-of-life impacts of CIDP (especially in the physical domain) and diagnostic delays exacerbate treatment dissatisfaction and loss of productivity, and they also contribute to substantial costs and medical resource use among patients with CIDP.**
- **Lack of benefit was reported by both physicians and patients, and poor tolerability was reported by patients as reasons for treatment dissatisfaction and discontinuation.**
- **These results highlight the need for additional or alternative treatments that provide improved health outcomes for patients.**
- **Due to limited long-term data, more longitudinal real-world evidence studies are needed to better understand the clinical, humanistic, and economic disease burden of patients with CIDP, especially for those who relapse or do not respond to initial therapy, and should also include caregiver burden and indirect costs.**

## REFERENCES

See supplementary material.

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## DISCLOSURES

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**SUPPLEMENT**

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